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Fibrous lesions of the extremities

Gerald A. Mandell, MD, H. Theodore Harcke, MD, and S. Jay Kumar, MD

FIBROXANTHOMA (NONOSSIFYING FIBROMA) AND BENIGN CORTICAL LESIONS OF THE DISTAL FEMUR

Fibroxanthoma, nonossifying fibroma, and fibrous cortical defect are all terms used to describe histologically similar lesions that occur in the growing bones of healthy children and adolescents between 2 and 20 years of age.[1] Thirty-three percent of the pediatric population has one or more of these lesions.[2] Most fibroxanthomas are detected as incidental radiographic findings.

The benign cortical irregularity or cortical desmoid of the distal medial femur seen in children and adolescents is a fibrous lesion that develops near the insertion of the tendon of the adductor magnus muscle. This irregularity, noted in approximately 36% of normal children,[3] is not seen after epiphyseal closure. A distal anterior femoral metaphyseal defect is also a normal variant.[4]

Etiology and pathophysiology

Solitary nonossifying fibroma and fibrous cortical defect are the most common benign bone lesions in childhood, and are of no significance. An association of multiple nonossifying fibromas in children with café-au-lait spots (Jaffe-Campanacci syndrome) has been described. Besides the cutaneous and osseous lesions in these patients, there are extraskeletal congenital abnormalities, which include mental retardation, hypogonadism or cryptorchidism, ocular anomalies, and cardiovascular malformations.[5–7] An increased incidence of the fibroxanthomas in patients with neurofibromatosis has also been described.[8–11]

The osseous lesions in neurofibromatosis may represent invasion by intraosseous neurofibroma, direct erosion by an adjacent lesion, or mesodermal dysplasia without associated neurofibromas.[10] In a recent report of a large series of patients without neurofibromatosis, the incidence of multiple lesions was 8%. Five percent of the patients with multiple lesions had neurofibromatosis.[12]

The term *fibroxanthoma* provides a very appropriate description of the underlying pathology. The lesion consists of spindle-shaped fibroblasts, scattered giant cells, and foam (xanthoma) cells. These lesions may ossify and become sclerotic. At this time, the term *ossifying nonossifying fibroma* has been used. However, the lesions in neurofibromatosis are usually large or even exuberant.[8]

The avulsive cortical irregularity of the distal femur is also referred to as superiosteal desmoid, subperiosteal abrasion, cortical abrasion, and distal metaphyseal femoral irregularity.[13] The male-to-female incidence ratio is 3:1. The lesion occurs twice as often in the left femur as in the right, and is bilateral in 25% to 35% of cases. Macroscopically, there is periosteal cortical thickening. Microscopically, there is evidence of fibrous tissue proliferation and numerous osteoclasts. The periosteal reaction, cortical thickening, reactive bone formation, and bony fragments within the soft tissues may be confused with malignancy. The pathogenesis seems to be related to mechanical stress at the point of insertion of the adductor magnus. This results in microavulsions, a disparity between bone resorption and formation, hypervascularity, and a fibroblastic response. The distal anterior femoral metaphyseal defect has an unknown etiology. The portion of the femur involved is intra-articular and probably does not represent an avulsion.[4] Keats and Joyce, however, believe that in adolescents metaphyseal

cortical irregularities can occur in other bones (such as the proximal humeral metaphysis, the upper humeral notch, or distal fibula). The radiologic similarity of these lesions suggests a common etiology, such as a variation of growth.[14]

Imaging

Recommended Approach

1. **Plain films.**
2. **Bone scintigraphy, to establish if a benign lesion has been traumatized and, if needed, to confirm the benign cortical irregularity.**

Plain radiography

Small lytic lesions with scalloped margins that occur eccentrically in a metaphysis and disappear spontaneously are termed fibrous cortical defects (Fig 1A). These lesions sometimes look like a blister with a very thin outer cortex. Larger, persistent lytic lesions with sclerotic margination that occur eccentrically but show interval growth are usually referred to as nonossifying fibromas.

The distal femoral cortical irregularity characteristically occurs in the posterior medial aspect of the bone, along the medial supracondylar ridge of the linea aspera femoris, just above the adductor tubercle at the insertion of the adductor magnus. The external oblique anteroposterior (AP) radiograph demonstrates it well.[13] Measuring approximately 1 cm to 3 cm in length, it can appear as a lytic area with periosteal reaction and sometimes spiculation (small cortical fragments in the associated soft tissue). These features raise the suspicion of malignancy. Biopsying the lesion is not helpful because the histologic appearance may result in a false-positive diagnosis of malignancy (most commonly osteosarcoma or fibrosarcoma). The distal anterior femoral metaphyseal defect is seen in the lateral radiograph as a defect in the cortex of the anterior aspect of the distal femoral metaphysis, just above the growth plate.[4]

Bone scintigraphy

The nonossifying fibromas (fibroxanthomas) and generally smaller fibrous cortical defects can show normal or increased uptake (Fig 1B).[1,15] Mildly active eccentric foci can be seen in the metaphyses (fibrous cortical defects) or the metaphyseal-diaphyseal locations (fibroxanthomas of the long bones in the lower extremities) (Fig 2). Normal scans are obtained when the lesion is quiescent.[15]

A more marked increase in activity can be physiologic (conversion from nonossifying to ossifying fibroma) or posttraumatic (pathologic fracture). The ability to detect uncomplicated lesions also depends on size of the lesion, and the resolution of the imaging equipment in addition to the osteogenic activity.

The benign cortical irregularity has normal or slightly increased uptake on bone scan.[6,16] In contrast, an osteosarcoma would have a marked increase in the amount of activity at the tumor site. When the patient presents with pain, the bone scan can very easily differentiate this benign lesion from the malignant alternative.

Magnetic resonance imaging

The MR appearance, similar to the plain radiographic characteristics of fibrous lesions, is quite variable depending on the status of the lesion. Early developing lesions demonstrate high signal intensities of varying homogeneity on T1-weighted images.[17] The high signal is consistent with varying content of fat-storing foam cells. On T2-weighted images, the signal intensity reduced secondary to the intracellular fatty substances stored in foam cells and the intracellular hemosiderin of stromal cells. Low-signal linear structures correspond to fibrous septa and osseous pseudosepta. With progressive ossification or maturation, the diaphyseal portions of older lesions exhibit lower signal intensity on T1-weighted images. During further healing, there is the appearance from the diaphyseal side of the lesion of the density of yellow bone marrow (Fig 3). Some lesions can have mixed signal characteristics; the more diaphyseal portion produces less signal and the paraepiphyseal a brighter signal on T1-weighted images. Sometimes, on both T1- and T2-weighted images portions of the lesion display no or decreased signal. Alteration of the signal may be attributable to the deposition of hemosiderin and the larger presence of collagen.[18]

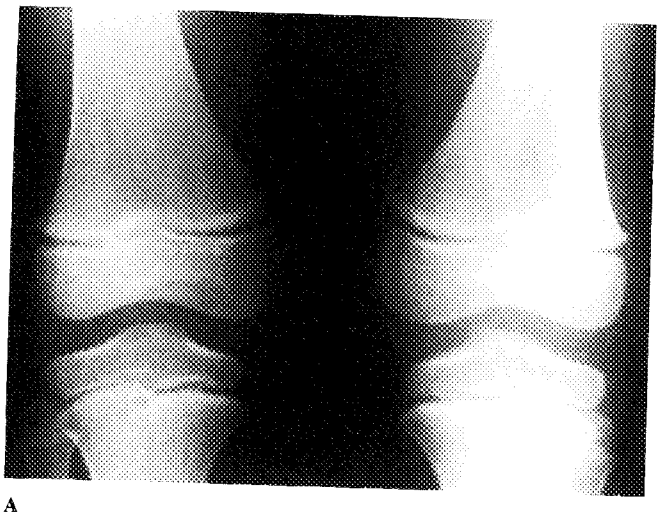
Clinical course

Nonossifying fibromas and fibrous cortical defects are typically asymptomatic. Occasionally, pathologic fractures result at this point of cortical weakening.[19] The benign cortical defects, which have very thin cortices, may be most prone to fracture. A muscle pull can produce an avulsion fracture, which subsequently induces periosteal reaction.[19]

Sometimes patients with benign cortical irregularities of the distal femur present with knee pain. Eventually these lesions resolve spontaneously. It is important to perform bone scintigraphy when clinical and radiologic differentiation of benign lesions from malignancy is necessary.

Differential diagnosis

Cortical desmoid, intracortical fibrous dysplasia (ossifying fibroma of long bone), and cortical chondromas must be distinguished from small fibroxanthomas or fibrous cortical defects. The larger intramedullary fibroxanthoma must be differentiated from chondromyxoid fibroma, brown tumors of hyperparathyroidism, and fibrous dysplasia. If there is no clinical evidence of hyperparathyroidism, then brown tumors are unlikely. Chondromyxoid fibroma usually has more sclerotic, scalloped margins and occurs predominantly in the proximal tibia and in the feet. The lesions of fibrous



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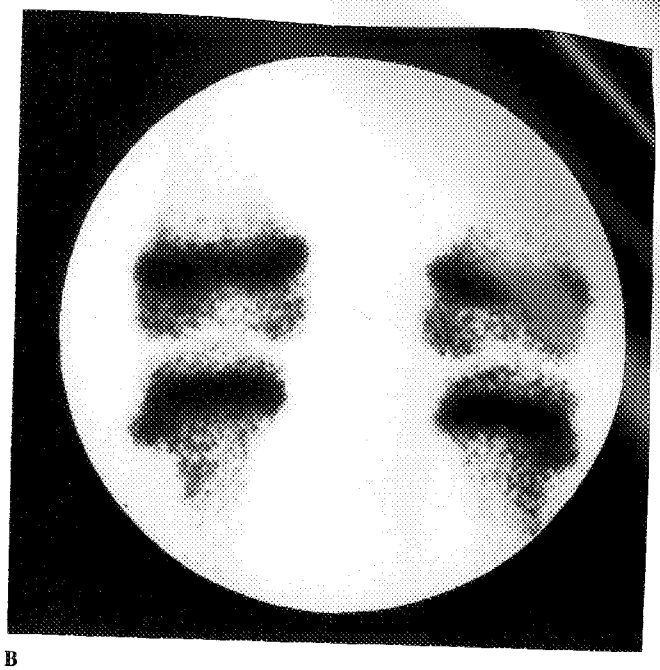

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Fig 1. Cortical desmoids in a 7-year-old with knee pain. **(A)** Radiograph: AP view of the knees reveals areas of lysis in each medial metaphysis. **(B)** Bone scan: Anterior image of the knees shows slight physiologic activity in the medial metaphysis.

dysplasia are extremely active on bone scintigraphy. The osteofibrous dysplasia (ossifying fibroma of the long bone) is usually located in the mid-tibia and associated with tibial bowing.

Treatment

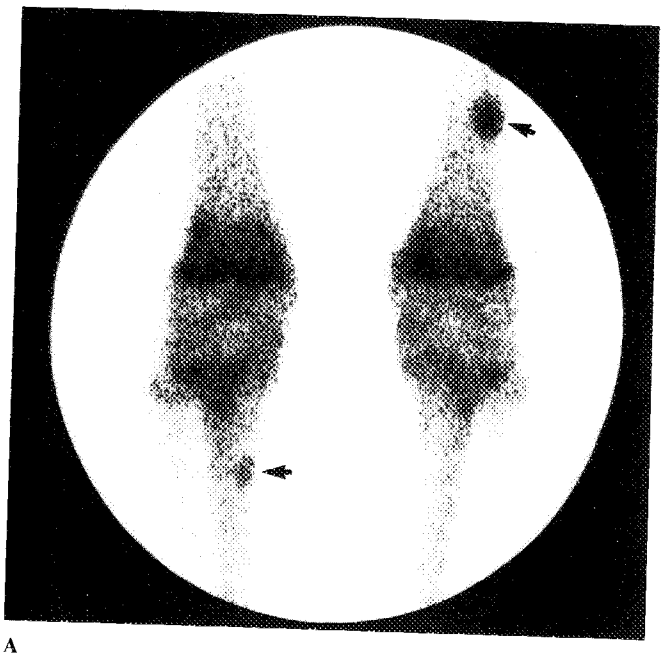
No treatment is necessary for uncomplicated lesions.

DESMOPLASTIC FIBROMA

Desmoplastic fibroma is a locally aggressive bone tumor that morphologically resembles the soft-tissue desmoid tumor of the abdominal wall. It is a rare and benign tumor.

Etiology and pathogenesis

The tumor consists of small, benign-appearing fibroblasts in a matrix of collagen. Mitotic figures are rarely observed. The lesion is identical to the desmoid tumor found in the abdominal wall and also to the periosteal or cortical desmoid seen in the metaphyses of the distal femur of children.[20] At times such tumors may be difficult to differentiate histologically from low-grade fibrosarcomas. The tumor is locally aggressive. Some



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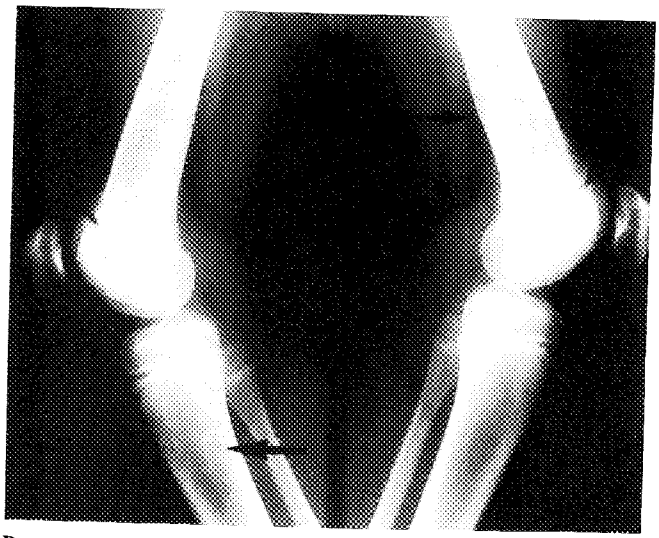

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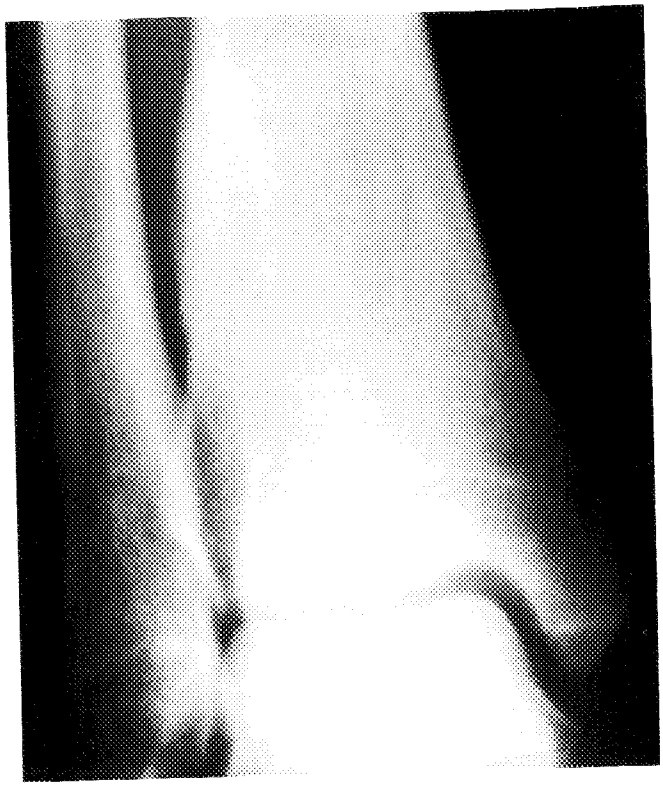
Fig 2. Fibrous lesions. **(A)** Bone scan: Anterior image shows multiple foci of increased uptake (arrows). **(B)** Radiograph: Lateral views correlate with the bone scan in **A** (arrows).



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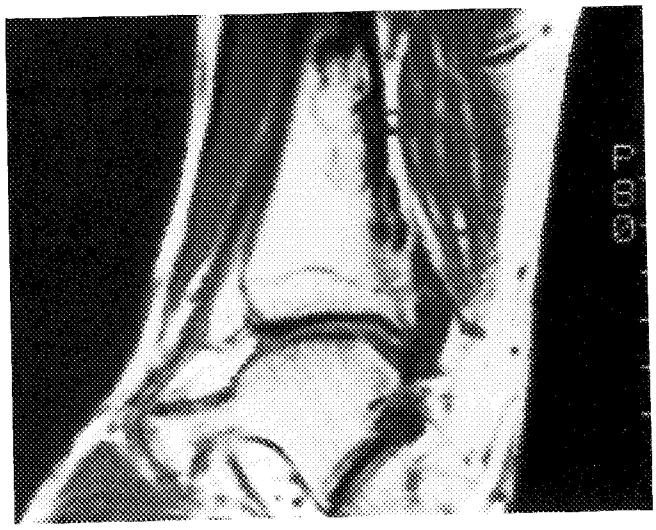

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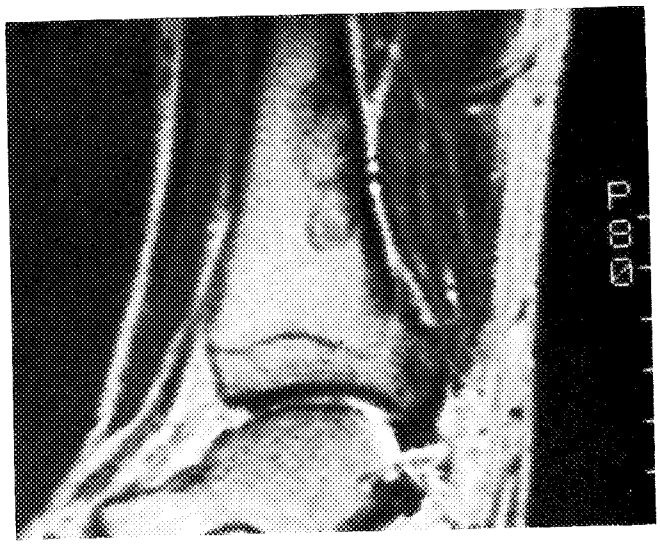


C

Fig 3. Fibroma in an asymptomatic 21-year-old patient. **(A)** Radiograph: AP view reveals the irregular cortical margin at the diaphyseal-metaphyseal junction of the lateral aspect of the tibia. **(B)** MRI: Sagittal T1-weighted (short-TR) image of the ankle shows a lobulated, eccentric lesion with internal signal density similar to the surrounding marrow, indicating a mature lesion. **(C)** MRI: In this sagittal T2-weighted (long-TR) image, fatty marrow causes a decreased signal both internal and external to the lesion. Note margination from the sclerotic rim.

lesions are hypovascular, others are hypervascular. These neoplasms have been found to occur from ages 20 months to 71 years, and most occur during the first three decades. There is no sex predilection. The lesion usually resides in the metaphysis of the long bone and sometimes crosses the epiphysis. Occasionally there is diaphyseal involvement. Most patients have a history of trauma.[21]

Imaging

Recommended Approach

1. **Plain films.**
2. **MRI.**

Plain radiography

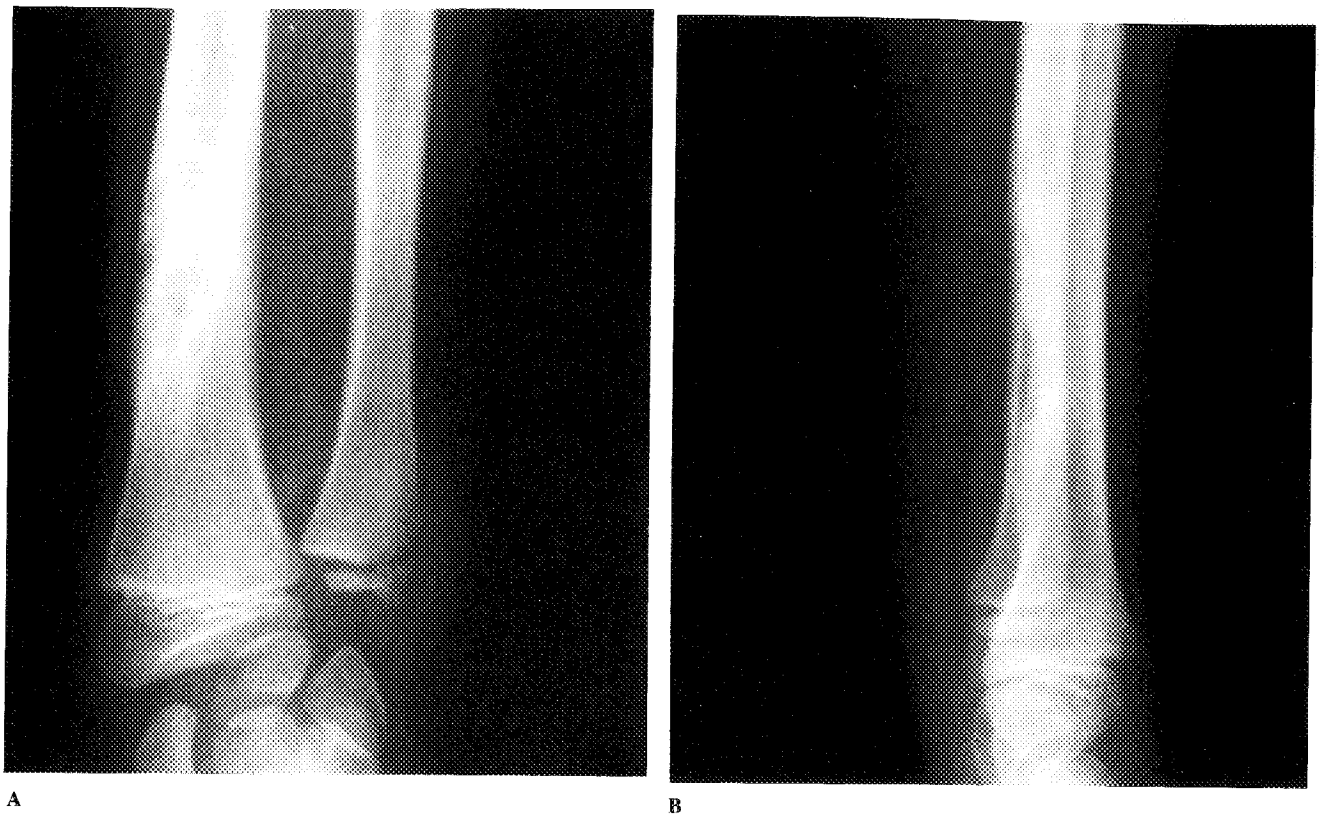
Radiographically, the tumor appears as an expansile radiolucent lesion with well-defined margins and destruction of the overlying cortex. There is extension into the soft tissues. Sometimes the tumor has internal trabeculation and exhibits a "soap-bubble" configuration. Usually there is no significant periosteal reaction. In one large series, the cortex was eroded in 28% of patients.[22] Cortical thickening is caused by apposition of periosteum (Figs 4A and 4B).

Bone scintigraphy

The few reported cases of desmoplastic fibroma show increased uptake on bone scintigraphy.[21,23]

Computed tomography

The lesion is well circumscribed, with a lobulated or scalloped margin, extensive cortical bone loss, and a well-defined associated soft-tissue mass. It is an eccentric, destructive lesion, with an intact inner margin and a variable amount of cortical bone loss (Fig 4C). CT can demonstrate soft tissue extension by the tumor.[24]



A B C

Fig 4. Desmoplastic fibroma in a 10-year-old patient. **(A)** Radiograph: AP view shows sclerosis in the distal diaphyseal-metaphyseal region of the left radius. **(B)** Radiograph: Lateral view pinpoints an area of sclerotic reaction and erosion on the ventral surface of the distal left radius (arrow). **(C)** CT: Transaxial (bone window) image. Note the irregular, thickened cortex of the lesion.

Magnetic resonance imaging

In an article by Crim et al, MR examination of one patient's distal femur was discussed. On T1-weighted images, the tumor was intermediate in signal density. The lesion was surrounded by a low-signal line, due to a pseudocapsule. On T2-weighted images, the tumor gave heterogeneous signal intensity, with persistence of the low-signal pseudocapsule.[22]

Clinical course

Pain and a palpable mass are common presenting symptoms. Nine percent of patients have a pathologic fracture.[22] Metastases have not been reported.[25]

Differential diagnosis

The desmoplastic fibroma can simulate a simple cyst, aneurysmal bone cyst, chondromyxoid fibroma, giant

cell tumor, fibrous dysplasia, or eosinophilic granuloma. Sometimes the trabeculation of the tumor can mimic a hemangioma.[20]

Treatment

Early curettage and bone grafting have resulted in a 40% rate of recurrence. Most surgeons try a wide resection with bone grafting. The histologic findings in recurrent tumors are the same as in the original tumor.

FIBROUS DYSPLASIA

Fibrous dysplasia is a benign, noninherited, developmental abnormality affecting the skeleton. The condition can be either mono- or polyostotic. There is a slight prevalence of females with the polyostotic form. Patients are often first seen during the first two decades of life. Such patients generally have pathological fractures and no other deformities. The osseous lesions and their association with skin pigmentation changes and dysfunction of the endocrine system were first described by Albright et al in 1937.[26] In one series, 66% of extremity lesions involved the lower extremities. Most foci were in the proximal femur.[27]

Etiology and pathophysiology

Most patients are recognized clinically in the first two decades of life. The appearance of the disease at an early age supports the hypothesis that it has a congenital origin. The monostotic variety is more common, and 75% of the lesions occur (with approximately equal frequency) in the skull, femur, and ribs.[28] In the polyostotic form, lesions are more common in the long bones, especially in the femur and tibia. The disease usually progresses until puberty, and then it stabilizes. After puberty, however, the disease may progress, although at a slower rate. Inactive disease may be reactivated by pregnancy.[29] Intraosseous lipomas, which are described in older adults in femoral neck locations similar to the sites where fibrous dysplasia occurs, are felt by some to represent an end stage (cystic degeneration with fatty or fibrous replacement) of fibrous dysplasia.[30]

Histologically, the lesion is composed of fibroblasts and fine collagenous fibers arranged in bundles and interspersed with irregularly shaped trabeculae ("woven bone").[31] Fibrous dysplasia is characterized by poorly oriented osseous trabeculae and islands of cartilage. Large amounts of atypical cartilage can be found in fibrous dysplasia in the absence of a cartilaginous neoplasm. One case has been reported, however, of chondrosarcoma arising in fibrous dysplasia without a history of previous irradiation that could account for the malignant transformation.[32] Fibrous dysplasia can be classified as a developmental disease of bone because abnormal fibrous tissue replaces normal spongiosa and fills the normal medullary cavity of affected bones. This fibrous tissue contains trabeculae of poorly calcified primitive bone formed by osseous metaplasia.

Albright syndrome, the combination of multiple endocrine abnormalities, skin hyperpigmentation, and fibrous dysplasia of bone, has an unclear pathogenesis.[33] It is not inherited. The organs most often affected are regulated by trophic factors that modulate intracellular concentrations of cyclic adenosine monophosphate.[33] Recent evidence suggests autonomous hyperfunction of the affected glands and organs. Cherubism, a familial disorder of childhood characterized by bilateral and symmetric expansion of the mandible and maxillary bones, has histologic and radiographic changes similar to fibrous dysplasia.[34] Congenital pseudarthrosis, characterized by fibrous degeneration of a segment of bone, is a condition that usually occurs in the tibia. The etiology of this disorder is uncertain, but it has histologic features that resemble fibrous dysplasia. This form of congenital pseudarthrosis must be differentiated from the pseudarthrosis seen in patients with neurofibromatosis.

Imaging

Recommended Approach

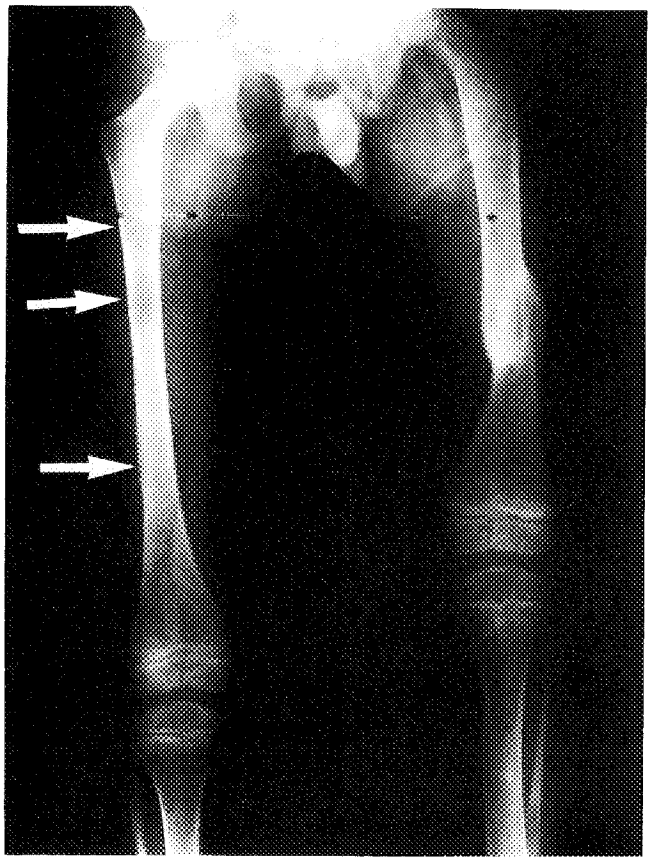
1. **Plain films.**
2. **Bone scintigraphy, to screen for multiple sites (then obtain radiographic correlation).**

Plain radiography

The characteristic roentgenographic changes of fibrous dysplasia are expansile, bubbly-appearing lesions accompanied by endosteal scalloping, bone deformity, and a hazy, or "ground-glass," texture. Radiographic density can range from purely lytic to classic ground-glass to sclerotic or densely calcified. There is usually associated overgrowth of the affected bone secondary to the chronic hyperemia that is part of fibrous dysplasia.

Fibrous dysplasia is categorized as monostotic or polyostotic, so it is important to evaluate the entire skeleton. There can be a solitary site or multiple sites in a single bone. A curious (and inexplicable feature) is the occurrence of dysplasia in several bones of a single extremity or the involvement of multiple bones, all on one side of the body. Lesions may also be scattered throughout the skeleton.

In the skull, expansion and replacement tend to selectively involve the outer table, with thickening and sclerosis of the inner table. Dense, uniformly sclerotic, lesions are most common in the occipital and sphenoid bones and sometimes occur in the extremities. Thickened bony septa may divide a site into multiple variably sized cavities. The process is one in which woven bone is produced that decreases overall bone density. This produces the "ground-glass" appearance described in fibrous dysplasia (Fig 5A).[28] In long bones, the lesions are usually metadiaphyseal in location. The shepherd's crook deformity of the proximal femur is the most



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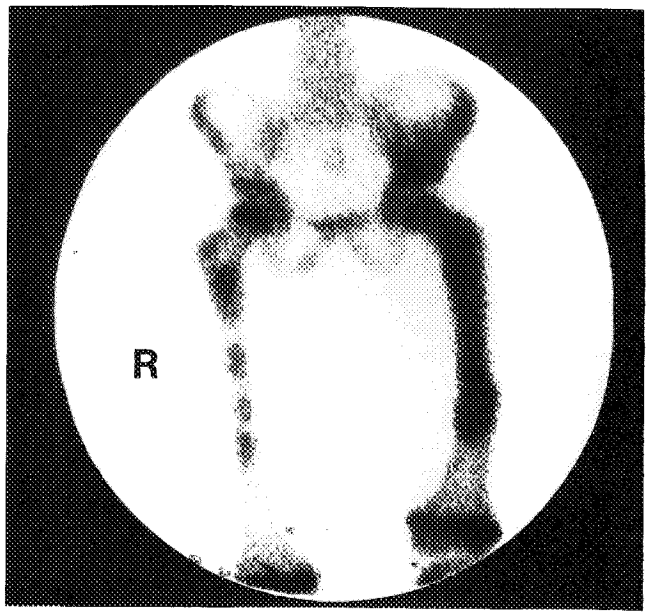

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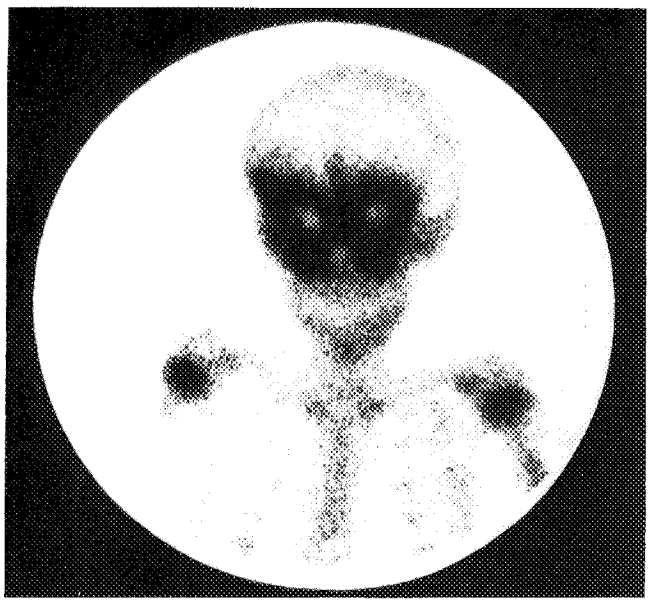

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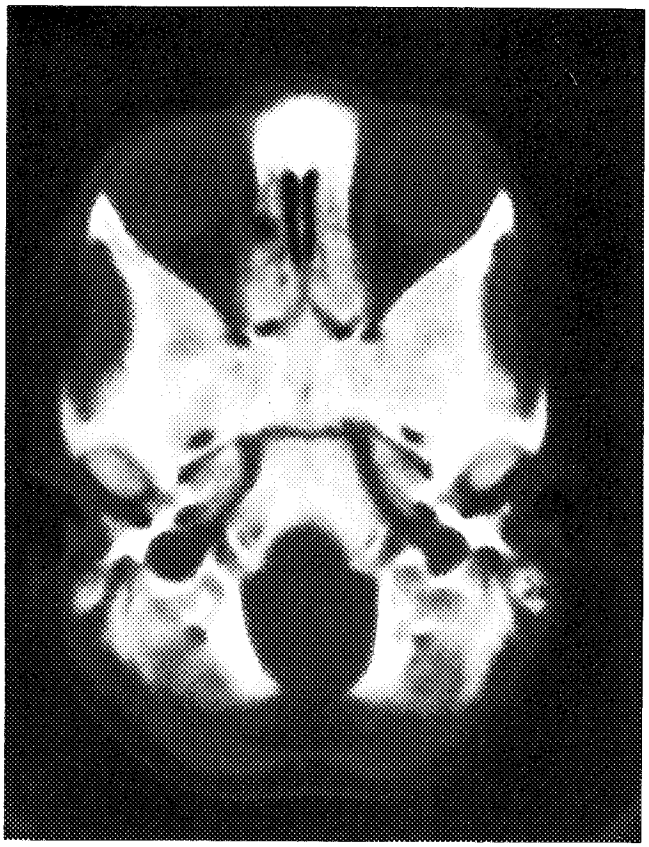

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Fig 5. Polyostotic fibrous dysplasia in a 6-year-old patient. **(A)** Radiograph: AP view of the femora shows the "ground-glass" appearance in multiple lesions in the right femur (arrows). The affected left femur has a healing fracture. Note the thin cortices. The left tibia is also involved. **(B)** Bone scan: Anterior images of the pelvis and femora reveal multiple lesions with increased uptake. Note healing left femoral fracture and shortening of the limb. **(C)** Bone scan: Anterior view of the skull. Formerly known as the "Lone Ranger" sign, the increased uptake around the orbits is now called the "Batman" appearance. **(D)** CT: This transaxial image (bone window) shows dense, thickened sphenoid bones. Note the posterior walls of the orbit.

common example of bone deformity associated with fibrous dysplasia. Vertebral lesions are rare.

Bone scintigraphy

Bone scintigraphy is a sensitive imaging technique for detecting early lesions and polyostotic involvement in fibrous dysplasia. Due to the increased vascularity of fibrous dysplasia, the lesions actively concentrate the bone-seeking radiopharmaceutical in both early perfusion phases and delayed bone phases of imaging (Figs 5B and 5C). In a report that studied 59 lesions, a small percentage of cystic lesions (14%) and homogeneous ground-glass lesions (7%) showed no increase in radioisotope uptake, although roentgenograms showed marked changes.[35] Bone scintigraphy, therefore, cannot be used alone to identify all sites of fibrous dysplasia. Typical locations that can be identified scintigraphically include (in decreasing order of incidence) ribs, tibia, femur, and maxilla (Fig 5B).[36] The orbital "mask" appearance (either monocular or binocular) is typical in fibrous dysplasia involving the facial bones (Fig 5C).

Pathologic fractures at a site of active fibrous dysplasia cannot be detected scintigraphically since there is avid concentration in the underlying lesion. When bone graft material is placed in an area of fibrous dysplasia, however, the activity detectable on bone scan can return to normal within a period of 6 to 12 months. If a pathologic fracture occurs in this situation in the area altered by surgery or at a site where the lesion is mature and more quiescent, then a linear focus of increased uptake can be recognized. High-resolution magnification views with a pin-hole collimator are sometimes helpful.

There is some debate as to whether cherubism is part of the spectrum of fibrous dysplasia. In one study, three of four patients with cherubism had normal bone scintigraphy of the jaws.[34]

Gallium-67 citrate has been reported to accumulate in regions of fibrous dysplasia. The mechanism by which a lesion attracts the gallium is not definitely known.[37] However, this finding prevents the use of gallium to detect malignant conversion of fibrous dysplasia.

Computed tomography

Plain films are usually sufficient for the diagnosis of most cases of fibrous dysplasia. CT has been reported to be highly useful for studying the extent of involvement in craniofacial lesions. Proptosis occurs in as many as 35% of patients with fibrous dysplasia. CT has been used to assess the activity of the disease by taking density measurements on lesions. Conversion from fibrous marrow to fibrous bone results in increased density measurements.

CT can demonstrate expansile, lytic, and well-defined lesions, as well as hazy, amorphous lesions. CT has been used in the differential diagnosis of lesions that mimic fibrous dysplasia because fibrous dysplasia usually has a much higher density (70 to 130 Hounsfield units) (Fig 5D). Lesions such as osteomyelitis, eosinophilic granuloma, and lytic neoplasms show lower density (20 to 40 Hounsfield units).[38] Occasionally, fibrous dysplasia produces soft tissue masses with extraosseous extension, and about 0.5% of fibrous dysplasia cases undergo malignant transformation. The malignant tumor types, in order of decreasing frequency, are osteosarcoma, fibrosarcoma, and chondrosarcoma.[31,32,39,40] CT or MRI can be used to assess the extent of soft-tissue involvement.

Magnetic resonance imaging

T1-Weighted images demonstrate a homogeneous decreased signal throughout the lesions.[41] T2-Weighted images have varying signal intensities. Most lesions exhibit a very intense signal on T2-weighted images. Some have inhomogeneity and septations. "Halo" lesions can also be seen, and feature central intermediate signal intensity surrounded by high signal intensity. MRI is very accurate at defining the extent of this osseous lesion.

Clinical course

The most common clinical presentation is pain and swelling in association with a pathologic fracture.[28] Eighty-five percent of patients with fibrous dysplasia develop pathologic fractures. A functional deficit such as a gait disturbance can follow as the patient grows or as the level of activity increases. Occasionally, a progressive cranial nerve deficit occurs secondary to foraminal narrowing about the face and skull. Alkaline phosphatase levels are usually elevated in patients with polyostotic fibrous dysplasia. Nonelevated brown or yellowish cutaneous macules with irregular borders can coexist with the osseous lesions in a third of patients with polyostotic fibrous dysplasia. The lesions are usually found on the trunk or buttocks, near the site of the bone lesion. A rare syndrome consisting of polyostotic fibrous dysplasia (in a predominantly unilateral distribution), cutaneous pigmented lesions, and precocious puberty is termed Albright syndrome. Other conditions that have been associated with fibrous dysplasia reportedly include hypophosphatemic rickets,[42] hyperparathyroidism, diabetes mellitus, arteriovenous fistulas, coarctation of the aorta, hypoplastic kidney, and soft tissue myxomas.[28] Whether the lesions of fibrous dysplasia eventually spontaneously resolve or become inactive with skeletal maturation or puberty is not well understood. After examining serial biopsy specimens, Harris et al reported only subtle histologic changes. No conversion of fibrous bone trabeculae to lamellar bone was noted.[27] Radiographic progression of disease, however, can occur 50% of the time, both before and after puberty.[27]

Malignant sarcomatous transformation can also occur.[32,38] Patients with the polyostotic type are more likely to develop a secondary malignancy.

Differential diagnosis

The radiographic differential diagnosis of monostotic fibrous dysplasia includes ruling out nonossifying fibroma, unicameral bone cyst, enchondroma, and giant cell tumor. In the differential diagnosis of polyostotic fibrous dysplasia, enchondromatosis (Ollier disease), brown tumor of hyperparathyroidism, histiocytosis X, and neurofibromatosis should be included.[28] In the older population, polyostotic disease can also be confused with Paget disease, osteoblastic metastases, or fractures.[36] Radiographically, fibrous dysplasia alters the normal outline of bone by its expansile nature, while with Paget disease the bone maintains its normal contour but the original osseous template becomes enlarged.

Treatment

Fractures in fibrous dysplasia are seldom displaced and usually can be treated with casting. Treatment results in satisfactory healing, particularly in non–weight-bearing bones. In the upper extremity, 88% of patients had satisfactory results following treatment of symptomatic lesions.[43] In the lower extremity, results are highly dependent on the patient's age at the time of initial presentation.

Fibrous dysplasia of the hip and the proximal part of the femur usually results in loss of structural integrity. The stresses of normal weight-bearing can produce pathological fractures.[29] In patients older than 18 years, the results obtained with closed reduction or curettage and bone grafting were satisfactory. In patients younger than 18 years, however, 88% of those with lesions treated with closed procedures and 81% of those treated by curettage and bone grafting had unsatisfactory results. However, 86% of lesions treated with internal fixation had a satisfactory result, independent of skeletal maturity and extent of disease (mono- or polyostotic).[33] Generally, monostotic hip disease is easier to control than polyostotic disease. Malignant transformation is usually recognized by an increase in pain at the involved site, often with associated soft tissue swelling.

In Albright syndrome, besides precocious puberty, certain endocrine hyperfunctions may appear, including hyperthyroidism, hypercortisolism, growth hormone excess, and hypophosphatemia.[33]

OSTEOFIBROUS DYSPLASIA (CAMPANACCI'S DISEASE OR INTRACORTICAL FIBROUS DYSPLASIA), WITH REFERENCE TO ADAMANTINOMA

Osteofibrous dysplasia or ossifying fibroma is a benign bone lesion that occurs most commonly in the tibia (92%) and occasionally in the fibula.[44] There is usually an associated tibial bowing in infants and young children with this lesion. Some cases previously reported as congenital pseudarthrosis of the tibia, as ossifying fibroma, or as congenital fibrous dysplasia may actually be cases of osteofibrous dysplasia.

Etiology and pathology

Osteofibrous dysplasia is a benign lesion that is often confused with fibrous dysplasia. The age at presentation ranges from birth to 20 years; six cases were diagnosed in the first 3 months of life. Sixty percent of children are younger than 5 years of age.[45] There is a slight male predominance.

The lesion was previously referred to as ossifying fibroma of long bone.[46] Histologically, the lesion consists of spicules of bone in a fibrous stroma, quite similar to fibrous dysplasia. The distinguishing features of osteofibrous dysplasia are zonal architecture, lamellar bone, and rimming of bone spicules by osteoblasts.[47]

Adamantinoma (ameloblastoma) can occur in association with either fibrous dysplasia or osteofibrous dysplasia.[48–50] Adamantinoma is an uncommon tumor, but 14% of these tumors do occur in children. Scattered epithelioid islands typical of adamantinoma can be found in an area of osteofibrous dysplasia.[50] This is explained by the concept that adamantinoma is a pluripotential neoplasm with a bimodal differentiation (epithelial and mesenchymal). The mesenchymal differentiation includes fibrous, fibrous-dysplasia–like, and osteofibrous dysplasia–like stroma. The proportions of each element are variable.[48]

Imaging

Recommended Approach

1. **Plain films.**
2. **CT.**

Plain radiography

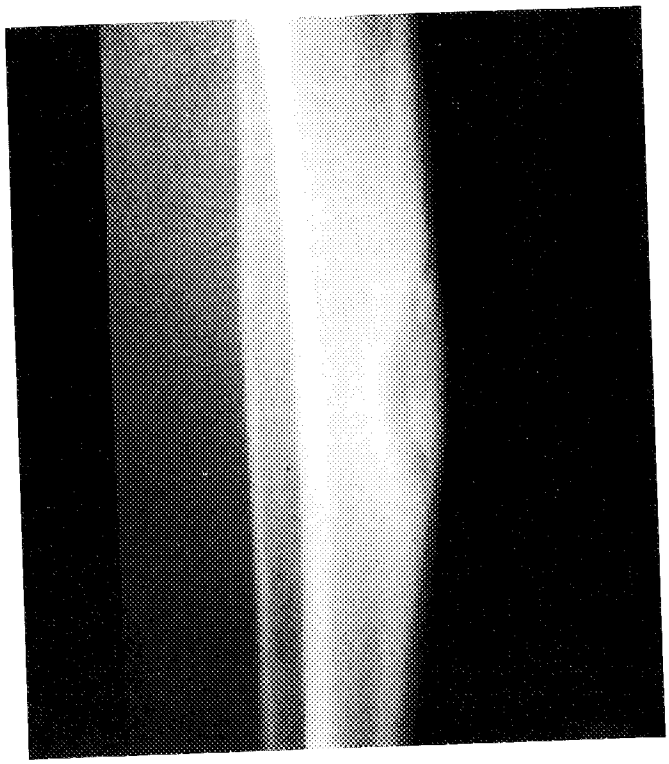
Seventy-one percent of tibial lesions involve the middle third of the bone. All fibular lesions are distally located. The radiographic patterns are age-related. In early infancy all lesions are unilocular, expansile, and lytic and have well-defined sclerotic margins (Fig 6A). The tibia appears bowed anteriorly. The lesion is usually found in the mid-tibia. After 3 months of age, the osteolytic lesion usually is eccentric and multilocular. The overlying cortex is thin and expanded, with a sclerotic rim along the medullary border (Fig 6B). The medullary cavity is usually narrow. If the lesion is large, the intracortical location may be radiographically difficult to define.

Bone scintigraphy

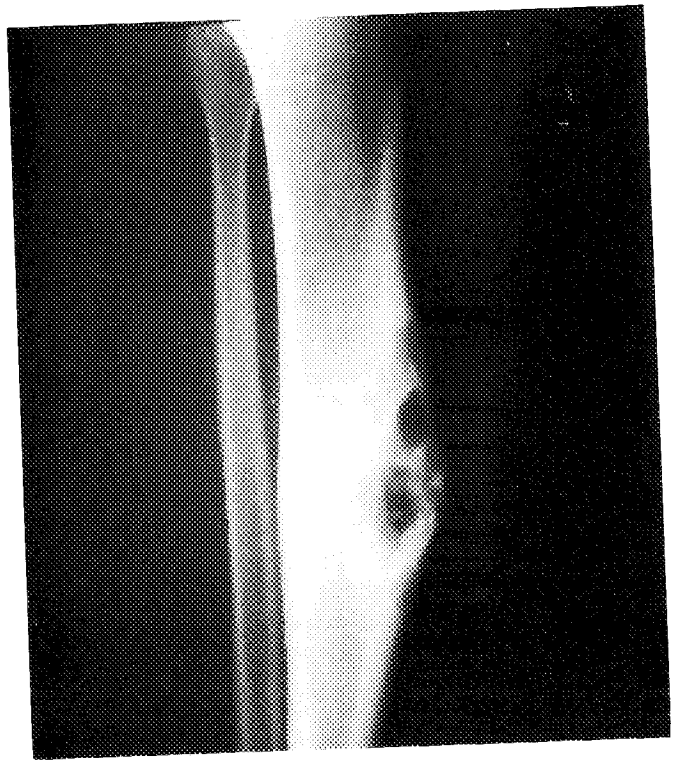
In our case and the one reported study, there was a marked increase of tracer uptake in the lesion.[51]

Computed tomography

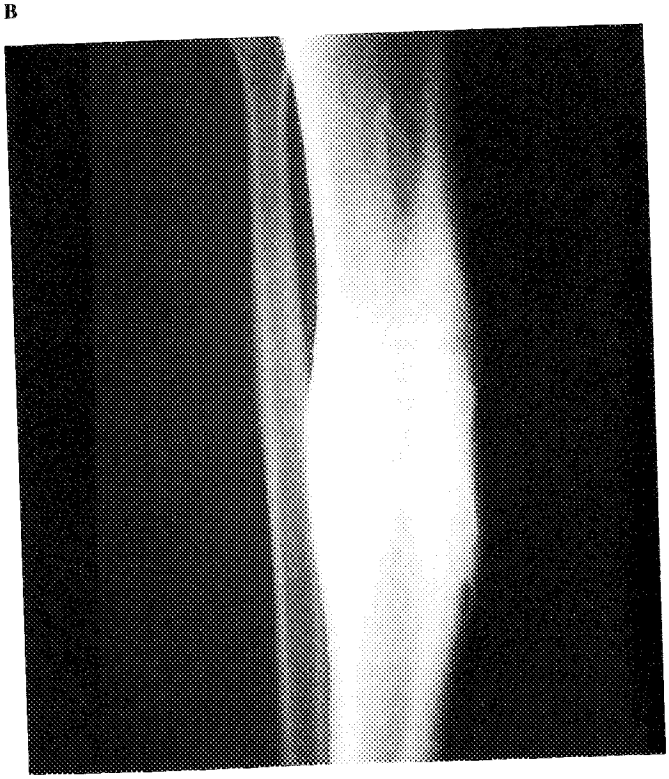
CT shows an intracortical lucent lesion with sclerosis of the inner margin. There is no associated soft tissue mass.[52]



A



B



C

Fig 6. Osteofibrous dysplasia (serial studies). **(A)** Radiograph: In a patient 8 years of age, a lytic lesion is beginning to form in the left anterior tibial cortex. **(B)** Radiograph: At 16 years of age, the polylobulated lytic lesion with sclerotic margination in the left tibia has increased in size. **(C)** Radiograph: By 17 years of age, the lesion is beginning to resolve and is developing some sclerotic thickening posteriorly.

Clinical course

Painless enlargement and bowing of the tibia occur. Rarely does a child present with a pathologic fracture.[53] The natural history is slow progression, and most untreated lesions stabilize or regress during childhood (after puberty) (Fig 6C). The lesion usually stops expanding after skeletal maturity is reached. Recurrence is unlikely after 15 years of age.

Differential diagnosis

Adamantinomas not associated with osteodysplasia can present radiographically as single or multiple radiolucencies with an admixture of sclerosis. The mid-tibial location and architecture make a solitary lesion difficult to distinguish from osteodysplasia.

Treatment

When there is unacceptable tibial bowing or pathologic fracture, surgery is necessary. Approximately 38% of the lesions may recur.[54] Observation is appropriate and some lesions may regress over time.

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